

Undifferentiated embryonal sarcoma of the liver mimicking a cystic mass

Konstantin Malley MD PhD*, Manvi Nagam MD, Nicco Buffolino, T. Brian Callister MD

Department of Internal Medicine, University of Nevada, Reno School of Medicine. *kmalley@med.unr.edu



University of Nevada, Reno
School of Medicine

Introduction

- Undifferentiated embryonal sarcoma of the liver (UESL) is an uncommon and aggressive neoplasm of mesenchymal origin occurring mainly in children age 6-10
- Cases in adults are exceedingly rare, with approximately 111 reports in the literature as of 2019¹
- Diagnosis can be missed due to lack of malignant or distinctive features
- A recent pooled analysis of UESL with 308 patients showed 5-year survival at 70% with partial hepatectomy, 79% with liver transplant, and only 6.6% with nonsurgical treatment only³

Case Description

- A 20-year-old male with no past medical history presented to the ER with a 5-day history of severe intermittent RUQ abdominal pain radiating to the shoulder, worse with movement.
- The patient had lived on a farm with goats and llamas as a teenager but never handled the animals. He had no history of international travel.
- Laboratory studies showed no evidence of biliary obstruction.
- Initial Abdominal CT read as abscess vs cyst
- Serum tumor markers CEA, AFP, and CA-19-9 were within normal limits



Figure 1. Arterial phase of 3-phase CT scan with non-enhancing mass in the right quadrant

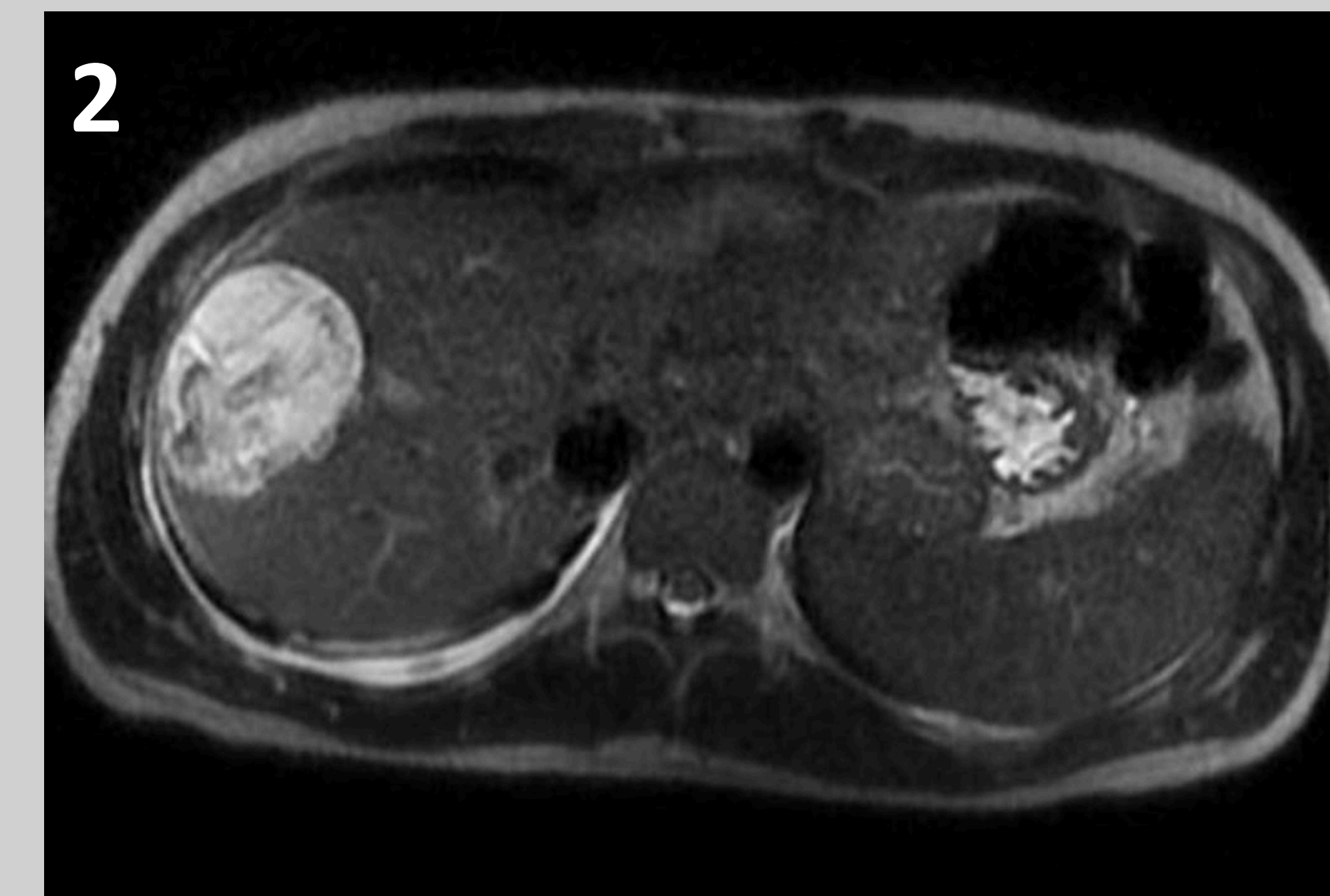


Figure 2: T2-weighted abdominal MRI demonstrating cystic features

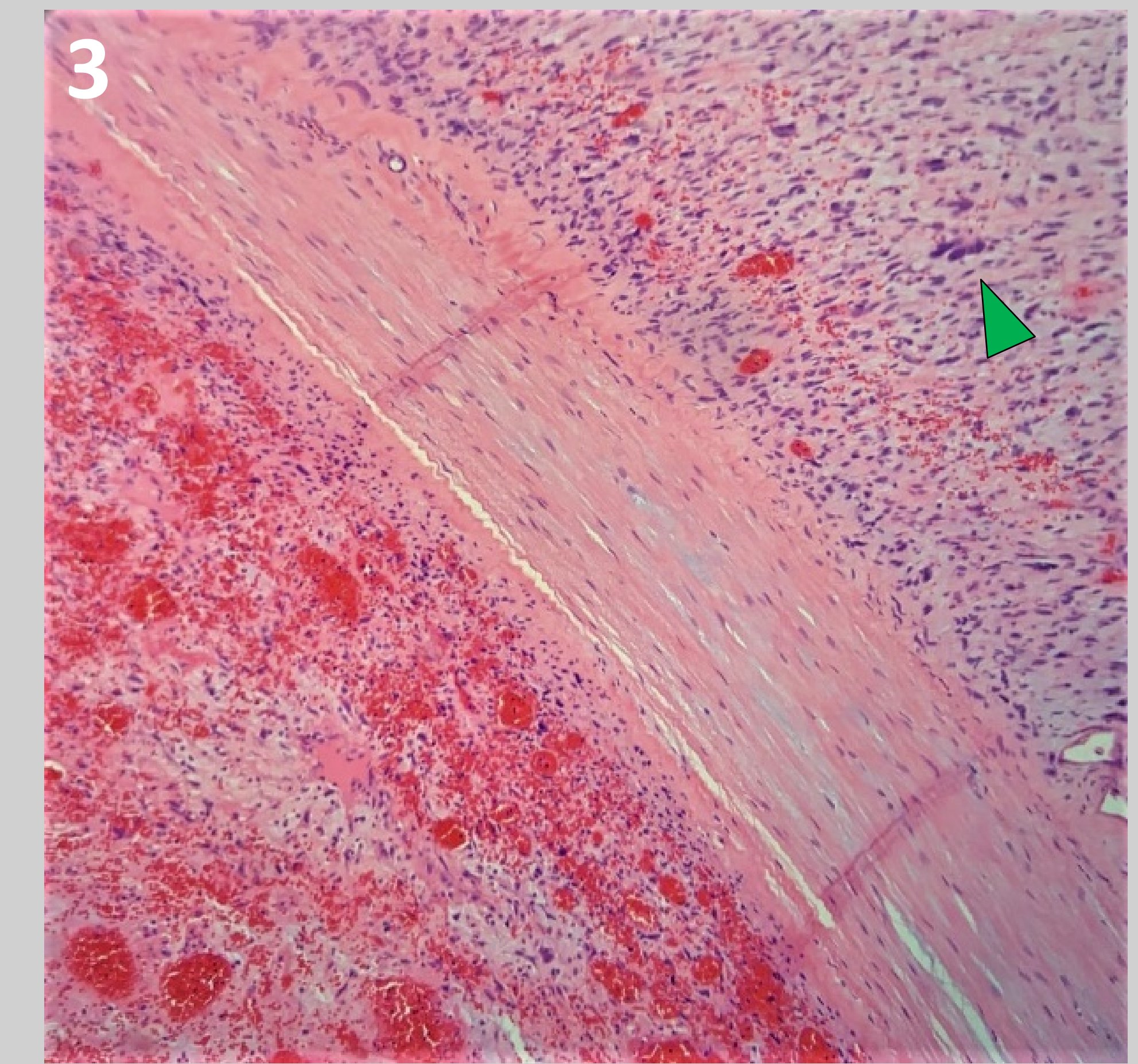


Figure 3: Liver mass core biopsy, H&E (10x), shows areas of hepatic parenchyma, fibrous stromal tissue, and hyperchromatic cells with surrounding mucinous tissue (green arrowhead)

Hospital Course

- On admission, IV ceftriaxone and metronidazole started to treat a potential abscess
- Radiology drained less than 1cc of thick, mucinous, blood-tinged fluid; core biopsy specimens obtained for pathology
- Hospital Day 2: Abdominal MRI showed possible cysts and no arterial enhancement or malignant features. Antibiotics discontinued and albendazole started to treat hydat
- Hospital Day 3: Discharged home in stable condition pending final pathology results. Serum IgG for echinococcus was negative.

Discussion

- Pathology showed atypical neoplastic cells with stellate forms. Positive stain for desmin, alpha-1 antitrypsin, glypican-3, among others together were consistent with UESL ; no single marker is diagnostic
- This case reinforces the importance of obtaining a tissue diagnosis as UESL can present with cystic features
- Discordance between imaging modalities (solid appearance on ultrasound, cystic on contrast studies) also should raise suspicion for malignancy and prompt a biopsy
- The patient recently underwent partial hepatectomy with microwave ablation, with tumor found adherent to diaphragm , complicated by abscess formation post-operatively

1. Shu B, Gong L, Huang X, Cao L, Yan Z, Yang S. Undifferentiated embryonal sarcoma of the liver in adults: Retrospective analysis of a case series and systematic review. *Oncol Lett.* 2020;20(4):102.
2. Neumayr A, Troia G, de Bernardis C, et al. Justified concern or exaggerated fear: the risk of anaphylaxis in percutaneous treatment of cystic echinococcosis-a systematic literature review. *PLoS Negl Trop Dis.* 2011;5(6):e1154.
3. Wu Z, Wei Y, Cai Z, Zhou Y. Long-term survival outcomes of undifferentiated embryonal sarcoma of the liver: a pooled analysis of 308 patients. *ANZ J Surg.* 2020;90(9):1615-1620.